THE ROYAL COLLEGE OF OPHTHALMOLOGISTS

Focus

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Giant cell - temporal - arteritis (GCA) in ophthalmic practice

Introduction:

Giant cell- temporal- arteritis (GCA) is a systemic vasculitis of the elderly that affects large and medium sized arteries with internal elastic lamina. It commonly affects branches of carotid circulation and hence is also known as cranial arteritis. Apart from the temporal arteries, this occlusive arteritis commonly involves the posterior cilliary arteries which can lead to severe visual loss by inducing ischaemic optic neuropathy. It is essential to establish prompt diagnosis and initiate treatment urgently as untreated arteritis can lead to progressive loss of vision and that of second eye involvement.

Clinical Features:

The varied clinical features of GCA reflect the systemic involvement, which can potentially involve any circulation. The clinical features can be described as systemic and ischaemic in nature. The systemic features often overlap with the related, milder inflammatory disorder- polymyalgia rheumatica. Proximal limb girdle weakness and stiffness, loss of appetite, weight loss and low grade fever may precede development of GCA. The onset of new headache is often a presenting symptom. A temporal headache and temporal tenderness are presenting symptoms at first consultation. Tenderness to touch the scalp is also a classic symptom. The history taking should include enquiry about jaw claudication (masseter muscle claudication), occipital headache or neck pain (occipital artery involvement), facial pain or swelling (involvement of facial artery). Tongue claudication is less frequent. The claudication symptoms are suggestive of intermittent ischaemia; scalp necrosis is rare but is indicative of established ischaemia. It is important to recognise that up to 20% of patients may not have any systemic symptoms and would still have occult GCA.

Ophthlmological complications:

The commonest presentation of GCA in ophthalmic clinic is with anterior ischaemic optic neuropathy (AION). It may be preceded by other transient ischaemic symptoms, e.g. amaurosis fugax, photopsia, or purple vision or diplopia. The optic nerve head classically has pale swollen appearance not dissimilar to a cotton wool spot. Often a cotton wool spot like swelling may be seen spilling over from the optic disc in to adjoining retina. (Fig. 1A). Arteritic (a) AION is associated with sudden loss of vision and an afferent pupil defect and altitudinal field loss is usually found. Unlike non-arteritic (na) AION, the aAION is associated with more severe loss of vision, chalky white swelling of the optic disc, normal/any sized optic disc (unlike small disc with no cup in fellow eye in naAION) and other vascular anomalies (e.g. choroidal non filling on Fluorescein Angiography). These features can be used in clinical assessment to support the clinical diagnosis of GCA.

Rarely a non embolic central retinal artery occlusion may be found responsible for loss of vision. A branch retinal artery occlusion is unusual but cilio-retinal artery involvement has been described. Occasionally a retinal artery occlusion may coincide with AION (Fig. 1B). The simultaneous involvement of two circulations (retinal and posterior ciliary) is highly suggestive of GCA. Some patients may have cotton wool spots in the retina preceding the development of AION. (Fig.1C)

Ocular ischaemic syndrome is a rare complication which indicates more generalised arteritic involvement of ophthalmic artery. The clinical features include ocular inflammation, low intraocular pressure and retinal/optic nerve ischaemia associated with ocular pain and loss of vision.

Diplopia may be caused by direct involvement of extraocular muscles in the inflammatory process in the orbit or may be central in origin. Ischaemic cranial nerve palsies may have variable presentation and may include medical 3rd nerve palsy – partial or complete, 4th nerve palsy and/or 6th nerve palsy. Multiple cranial neuropathies in presence of other recognised systemic symptoms should be considered to be due to GCA unless proven otherwise.

Diagnosis:

Clinicians should consider GCA as a possible diagnosis in patients presenting with ischaemic complications in ophthalmic practice. Finding of an ischaemic sign(s) in

presence of classic systemic features adds weight to the clinical diagnosis of GCA.

To confirm diagnosis, urgent blood tests should be requested that include full blood count, ESR and C–reactive protein (CRP). These may show anaemia and /or throbocytosis, however raised ESR and raised CRP are commonly used markers for GCA disease activity. Both these tests are useful in monitoring disease activity and response, and may help adjustment of steroid treatment. Combination of these two tests offers better indication than the tests on individual basis. A small minority of patients would have normal ESR and/or CRP.

The conclusive confirmation of diagnosis can only be made by the 'gold standard' temporal artery biopsy. Where possible, a temporal artery biopsy should be performed as soon as possible and within two weeks of initiation of steroid treatment. Histological examination of biopsy specimen typically shows pan-arteritis with round cell infiltration affecting all layers and can lead to occlusion of the lumen. Multinucleated giant cells are often found in the specimen but are not necessary for the diagnosis. Healed arteritis and skip lesions may be responsible for a false negative biopsy result and hence a long biopsy specimen (>2cm). A second artery biopsy may be considered, however in presence of typical clinical features of GCA, steroid treatment should not be discontinued. The American College of Rheumatology criteria (1990) for clinical diagnosis of temporal arteritis requires at least 3 of the following 5 criteria to be present (sensitivity 93.5%, specificity 91.2%) :

1. Age of onset older than 50 years
2. New-onset headache or localized head pain
3. Temporal artery tenderness to palpation or reduced pulsation
4. Erythrocyte sedimentation rate (ESR) greater than 50 mm/h
5. Abnormal arterial biopsy (necrotizing vasculitis with granulomatous proliferation and infiltration)

There is emerging evidence that non-invasive imaging such as Colour Doppler, MRI or fluorine-18-fluorodeoxyglucose positron emission tomography (FDG-PET) may aid the diagnosis . Finding of choroidal ischaemia / shut down on fundus fluorescein angiogram in presence of aAION or CRA occlusion can support the diagnosis of GCA.

Management:

In patients, presenting with visual loss due to aAION, or retinal artery occlusion, high dose systemic steroids, typically 1-2mg/kg/day is used. It should be emphasised that the treatment is to prevent further complications of arteritis as reversal of ischaemic complication is very unusual, especially recovery of vision. In cases of visual loss e.g. with aAION or ischaemic neurological deficits, patient should be admitted for high dose steroids and close monitoring . Initial treatment in such cases can be with intravenous steroids (methylprednisolone 0.5 -1g/day for up to 3 days). Apart from anti-inflammatory- immunosuppressive effect, it may also help with chemical decompression of optic disc oedema in cases of AION. Intravenous treatment should be followed by high dose oral steroids (Prednisolone 1-2mg/kg/day).The oral steroids need to be continued up to 2 years and in some cases indefinitely. The dose reduction needs to be gradual and titrated against clinical response as well as ESR and CRP levels. Along with steroids, protection for gastric mucosa (H2 blocker) and against osteoporosis (calcium supplements, bisphosphonates) should be considered. Usual precautions should be taken regards to pre-existing systemic infections prior to commencing steroids. Maintenance dose (5-10mg) of systemic steroids is usually required for up to 2 years for the majority of the patients. Relapses are rare with adequate treatment but need to be identified and treated promptly by adjusting to a higher steroid dose.

Some patients need second line treatment to reduce steroid dose and to control disease activity. Steroid sparing agents like azathioprine, methotraxate, cyclosporine, infliximab etc can be used however it is advisable that such patients are co-managed with rheumatologists.

Conclusion:

A high index of suspicion is needed for identifying GCA in patients presenting with visual disturbance suggestive of vascular origin. Clinical spectrum of GCA is varied but careful and systematic clinical evaluation supported by ancillary test can help to establish accurate diagnosis. Prompt treatment with high doses systemic steroids should be initiated, pending confirmation of diagnosis especially in cases of visual loss. Steroid treatment needs to be continued for long term and anticipated side effects of treatment should be discussed with the patient and appropriate preventive treatment should be offered.

Box 1: Clinical pearls

Clinical assessment: pointers to GCA
Systemic symptoms – Jaw claudication, Temporal artery abnormality
Severe vision loss
Chalky white disc
Other circulatory disturbance – e.g. diplopia/CRA occlusion
Investigations:
ESR/ CRP level – usually high but beware 'normal ESR GCA'
FFA – choroidal ischaemia in presence of AION or CRAO
TA biopsy – earlier the better, use Doppler to mark the course of artery.
Treatment:
Treat early treat hard- consider IV steroids when presenting symptoms are severe
Protection for gastric mucosa, bones
Collaborate with Rheumatologists
Second line agent
Long term care

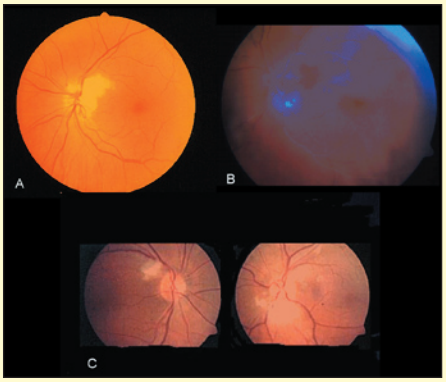

Fig.1. Ophthalmic complications of GCA

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